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ABSTRACT

This paper describes characteristics of and educational implications for children with Hunter's syndrome, a rare, genetic lysomal storage disorder resulting from an absence of the enzyme iduronate-2-sulphatase. Boys born with this sex-linked disease are born with little or no clinical manifestations, but generally are diagnosed by the age of three due to developmental delays. This progressive disorder results in early mortality and presents a myriad of considerations for special education. The paper addresses the history of the disability and its symptoms, including cognitive disabilities, speech development, eating problems, ear infections, and respiratory problems associated with the disorder. It notes that progressive physical and cognitive disabilities require special educators to work with a team of professionals to provide these children an appropriate education and a better quality of life. Behavior problems of the children with Hunter's syndrome are also described and include hyperactivity and aggressive behavior. Strategies are provided for encouraging social relations among students with disabilities and their typical peers. The importance of parents as part of the collaborative team to ensure successful education of these children is stressed. (Contains 17 references.) (CR)

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Hunter's Syndrome : Description and Educational Considerations

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Abstract

Hunter's Syndrome is a rare, genetic lysosomal storage disorder, resulting from an absence of the enzyme iduronate-2-sulphatase. Boys born with this sex-linked disease are born with little or no clinical manifestations, but generally are diagnosed by the age of three due to developmental delays. This progressive disorder results in early mortality and presents a myriad of considerations for the special educator. Progressive physical and cognitive disabilities in these children offer the special educator a chance to work with a team of professionals to provide these children an appropriate education and a better quality of life. Behavioral interventions can help children gain more control over their hyperactive and aggressive behavior. As part of the collaborative teaming of professionals, parents are vital to the successful education of these children. Collaborating with parents is essential to a successful program.

Hunter's Syndrome : A Description and Educational Considerations

Hunter's Syndrome, type A is a rare disease, which leads to multiple impairments and early death (Young and Harper,1983). Although this type of disability is low-incidence, teachers can learn effective ways to educate students with Hunter's in the least restrictive environment (Lewis and Doorlag,1995). With the passage of special education laws, special educators and general educators will have physically, behaviorally, and cognitively disabled children in their classrooms (Shaughnessy,1996). In addition to educating these children, teachers will be working with members of the collaborating team who are specialized in specific areas. These team members will work along with teachers to educate, and assist these students. Parents are also a part of the collaborating team.

Teachers need to become sympathetic to the needs of the families, so that the families become involved in their children's education (Simpson, 1996). Becoming familiar with all aspects of Hunter's syndrome, learning methods to assist the education of children with this disease, and collaborating with other professionals and parents will increase the quality of the children's education, and life.

History and Description of Hunter's Syndrome

Hunter's syndrome is one type of lysosomal storage disorders known as mucopolysaccharidoses (Muenzer, 1986). The mucopolysaccharidoses (MPS) are metabolic diseases caused by deficiencies of enzymes which normally would degrade mucopolysaccharides. Mucopolysaccharides are complex sugars which are made up of dermatan sulphate and heparan sulphate. These occur naturally in people and are broken down and excreted from the body except in the absence or mutation of the lysosomal enzyme iduronate-2-sulphatase(IDS) structural gene. In the case of the absence or mutation of IDS, dermatan sulphate and heparan sulphate accumulate in many tissues including organs, connective tissue and the central nervous system (CNS) causing many physical, behavioral and cognitive disabilities.

There are many different types of MPS diseases resulting from a variety of missing lysosomal enzymes. In addition, there are degrees of severity according to the absence or mutation of the enzymes (Wehnert, Hopwood, Schroder & Herrmman, 1991, Wraith, Cooper, Thornley, Wilson, Nelson, Morris & Hopwood 1990). MPS II (Hunter's Syndrome) is a genetic disease that is X-linked recessive. MPS IIB is the mutation of the IDS enzyme (mild Hunter's). MPS IIA is the total absence or mutation of the enzyme (Severe Hunter's).

Hunter's syndrome affects males only and is genetically transmitted from mothers to their sons (Muenzer, 1986) Mothers are carriers and except in rare cases, females are not affected (MPS Society). Mothers have a 50 % chance of transmitting the IDS structural gene to their sons who would become affected and 50% chance of transmitting the IDS structural gene to their daughters who would become carriers. Many mothers of sons with severe Hunter's syndrome are not carriers of the affected gene. Even among severe Hunter's boys, there is a clinical variability.

Clinical Manifestations

Severe Hunter's syndrome is a progressive lysosomal storage disorder resulting in the accumulation of muccopolysaccharides in the CNS and peripheral tissues. The clinical results of this accumulation are the progressive degeneration of the CNS, musculoskeletal system, respiratory system, cardiovascular system, gastrointestinal system, and other systems. In all severe Hunter's children, death generally occurs between the ages of ten and fifteen
(Muenzer, 1986).

CNS manifestations include degrees of mental retardation, behavioral problems, moderate to severe difficulty in communicating, hydocephalus, and seizures
(Young and Harper, 1983). In a study of 52 Hunter's patients, Young and Harper found a range in IQ's of less than 10 to 84. In all the severe Hunter's subjects, IQ dropped as the disease progressed. All generally begin to regress in cognitive ability at, or before the age of seven when the CNS becomes more involved. Children have limited and sometimes no speech. Also around the age of seven, and for the same reason, affected children will have some degree of communicating hydrocephalus (fluid in the brain) with elevated intracranial pressure. Some have had cognitive improvement with relief from a shunt which drains the fluid.

Behavioral problems include degrees of hyperactivity and variable destructive behavior (Young and Harper, 1981). These behaviors change around the age of seven when the children become generally quieter and less active. In the later stages of the disease, affected children become uncommunicative and immobile. At varying stages of the disease, but generally after the age of seven, it is common for children to have seizures.

The musculoskeletal manifestations give Hunter's children their characteristic appearance (Muenzer,1986). They are generally normal in appearance at birth. Around the age of two or three, changes in the facial appearance become apparent. Facial hair becomes increasingly coarse and thick. Eyebrows become thick and dark. The bridge of the nose becomes depressed and there is a progressive thickening of the nostrils, lips, and tongue (Shapiro, Stome, & Crocker, 1985). The tempero-mandibular joint or the jaw becomes stiff and painful resulting in poor jaw movement and reduced chewing of food.

As Hunter's syndrome progresses, perhaps because of limited use, muscles become atrophied and tight Achilles tendons are common (Young & Harper, 1983). Their arms and legs will increasingly retract, limiting their range of motion. They have limited rotation of their heads and wrists.

Respiratory problems are the leading cause of mortality in children with severe Hunter's syndrome (Shapiro, Stome, & Crocker, 1985). These are caused in part due to decreased mobility of the chest wall and deformities in the trachea and bronchial cartilages. Other problems result from upper airway obstruction, and obstructive airway disease. Early on, children have excessive mucous in their upper airway and recurrent rhinitis (nasal infections). A study of 31 cases show frequent upper respiratory infections, including tonsillitis, laryngitis, and otitis media. There are often thickened secretions in the nasal passages and trachea. The tonsils and adenoids are enlarged. Anesthetic procedures become difficult as a result. Sleep apnea (the cessation of breathing for a period of time) occurs in many cases. Recurrent bronchitis and pneumonia are common in the later stages of the disease.

Gastrointestinal manifestations in severe Hunter's syndrome include enlargement of the liver and spleen, chronic diarrhea, and inguinal and umbilical hernias (Young & Harper, 1983). Large livers and spleens are common in affected children resulting in protruding abdomens. Chronic and intermittent diarrhea is a common feature. Many children are treated early in the disease for inguinal and/or umbilical hernias. Some children with severe Hunter's syndrome have degrees of visual and/or hearing impairment caused by retinal detachment and recurrent otitis media
(Young and Harper, 1983). Deafness or hearing impairment is many times improved with the placement of tubes.

Educational Considerations

In the future, teachers can expect to have students with multiple disabilities (Shaughnessy, 1996). Children with severe Hunter's syndrome have multiple disabilities and fall into the category of other health impaired in the Individual with Disabilities Education Act (the IDEA, P.L. 101-476) (Lewis and Doorlag, 1995). They have degrees of mental retardation, behavior disorders, and physical disabilities which are progressive (with the exception of behavior which improves as the student's health deteriorates). This presents a myriad of considerations for the special educator.

In addition to the education of the affected child, there are issues of support and collaboration with parents of children with this disease (Simpson, 1983). With the collaboration of professionals, para professionals, teachers and parents, students with severe Hunter's syndrome can increase their quality of life by learning better communication, learning skills, having opportunities to integrate with peers, and learning to gain control of some of their inappropriate behaviors.

In the past, boys with Hunter's syndrome have been institutionalized (Young and Harper, 1983). In the 1960's, normalization took many of these individuals out of institutions and placed them back into the mainstream
(Simpson, 1983). P.L. 99-457 gave severely disabled students the right to a free and appropriate education. As services for this group of students evolved, they were taken out of institutions and placed in more" normal" environments. Due to the changes in education, students with severe Hunter's syndrome are entering the public schools. Awareness of the physical aspects of this disease, common behavior characteristics, and cognitive potential of these individuals will be helpful in planning for the educational needs of these students.

Physical Considerations

Special educators should individualize instruction for students with Hunter's syndrome and discuss any medical concerns with the parents and medical professionals on the collaborating team (Polloway and Patton, 1997). Medical and physical interventions should be planned according to school policy. Children with severe Hunter's syndrome have some common physical disabilities of varying degrees. Those physical disabilities that would be considered in a school setting are progressive : muscle contraction and atrophy, skeletal deformities and arthritis type pain, recurrent ear, nose and chest infections, sleep apnea (cessation of breathing when asleep) speech, and gastro-intestinal problems.

Musculoskeletal

Detection of Hunter's syndrome occurs in the first three years of life because children miss developmental milestones. Young and Harper (1983) report the following developmental delays in their study of 52 children. For 26 cases, the average age for sitting unsupported was 8.1 months. In 42 cases studied, the average age for walking was 15.7 months. Joints become progressively stiff. The fingers and hands become "clawed" or contracted. This makes fine motor skills increasingly difficult. Occupational therapists can help children gain skills, and with vigorous therapy limit the contractures to a degree. Using functional skills such as rolling out play-dough, or pushing doors open using the palms of their hands are examples of functional therapy (Shea & Bauer, 1994). Besides fine motor disabilities, children with severe Hunter's have gross motor impairment. The legs and arms become restricted in their movement. Physical therapists can provide activities that will stretch the muscles. Activities for stretching these muscles can be naturally occurring like jumping on a trampoline, and riding tricycles or other car type vehicles where their legs are fully extended at some point. This will delay the progression of muscle atrophy, and the contraction of large muscles. Passive stretching and bending of the joints can cause damage and pain (MPS Society). Boys with Hunter's are generally very active and love to play.

Respiratory

Recurrent chest infections are common in children with severe Hunter's syndrome (Shapiro, Stome, & Crocker,1985). As with the other tissues, mucopolysaccharides build up in the lungs, cartilage, and other areas of the lungs. The breastbone is not as flexible as it should be and the shape of the chest is abnormal. As a result of this, and large internal organs, the lungs have an increased amount of secretions which are hard to clear. Clearing the system of secretions becomes more difficult as the children age. They often have productive coughs. This becomes worse when they get bacterial infections or allergies. Parents, school nurses, and/or the family physician can help the collaborating team select the best posturing and intervention to help make them more comfortable. A physiotherapist can instruct teachers how to administer postural chest drainage.

As a result of respiratory involvement and trachea deformity, sleep apnea is common (Shapiro, et al, 1985). Another possibility for sleep apnea was suggested in a case study (Kurihara, Kumagai, Goto, Imai & Yagishita, 1992) Kuihara et al. described a boy who had frequent sleep apnea. Sleep studies conducted showed an average of 13.3 apnic episodes per hour for a total of 143 episodes in one night. Breathing stopped for a total of 37.4 minutes during the night. Autopsy showed storage material in the neurons of the respiratory center suggesting that the reason this child stopped breathing so often was of neurological origin. Regardless of origin, teachers need to take precautions. When these children take naps at school, they need to be positioned in such a way that their necks are as straight as possible. This will keep their airway clear. During activities, they can be encouraged to have good posture. Parents and medical professionals can show teachers what sleep position is best for their child.

In addition to excessive mucous in the lungs, children affected with severe Hunter's have recurrent ear, nose , and throat infections (Shapiro, et al., 1985). Even when they are not sick, they often have nasal drainage. They are noisy breathers. Teaching children to blow or wipe their own nose would be an appropriate and functional goal for daily living skills classes.

Ears

Ear infections are common in affected children (Muenzer, 1986). Young and Harper (1983) reported that 47 boys with Hunter's syndrome were suspected of having varying degrees of deafness. Formal hearing assessments were difficult or impossible due to behavioral problems. The data from hearing tests that were available showed both conductive and sensorineural deafness. The placement of tubes showed temporary improvement in hearing. Some children wear hearing aides. If this is the case, the teacher must be diligent in seeing that the hearing aides are not taken out and chewed. As with any student with hearing impairment, teacher and others can use good teaching practices to increase communication. Facing children, and using cued speech are helpful tactics in increasing children's receptive communication (Shea & Bauer, 1994)

Mouth

Eating can be difficult for children with severe Hunter's. Large protruding tongues and painful jaws can result in affected children not chewing their food (MPS Society). Storage of mucopolysaccharidess in the esophagus also complicates eating. The area for food in the esophagus in limited. As a result, choking is a problem for affected children. Teachers can take preventative measures to reduce the risk of choking. Food should be cut up into very small pieces. Soft food or pureed food is recommended when the child has problems chewing and/or swallowing .

Some affected children learn to use silverware and drink from cups. Depending on the level that they are at, they can learn to feed themselves as part of daily living skills classes. Offering choices of food is a good practice and will allow them the opportunity and motivation to use expressive language (MPS Society).

Speech

Young and Harper's (1983) study revealed data on speech development for 36 individuals with Hunter's syndrome. Of the 36, five never spoke meaningful words. The average age for speaking meaningful words for the other 31 individuals was 17 months. Of these 31 individuals, only five had spoken over two or three word sentences. They spoke in repetitive sentences, and exhibited " echolalia or mindless repetition of nursery rhymes" (p.484). There are some similarities in the speech patterns of children with Hunter's syndrome and autism.

The Indiana Resource Center for Autism lists perseveration and rambling as characteristics of children with autism (1979). Speech and language pathologists can work with individuals to increase their ability to articulate and make their needs known (Shea & Bauer, 1994). Using methods to develop their language concept development will help increase their receptive communication (Thomas, 1986). If they are not able to articulate, other forms of communication such as gestural, or aided systems can be considered.

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Gastrointestinal

Many children with severe Hunter's syndrome have recurring bouts of diarrhea (Young and Harper, 1983). In their study of Hunter's children, Young and Harper noted 22 out of 36 cases of diarrhea. Possibly related were the details of toilet training for 35 patients. Nineteen never gain control of bowel function and of the remaining sixteen, five never became dry at night. If control is gained, children become incontinent as the disease progresses. Inguinal and umbilical hernias are common. The collaborative team can determine learning priorities for children and decide if toilet training is an appropriate educational goal.

Neurological

One of the defining characteristics of severe Hunter's syndrome is the progressive neurological involvement (Young and Harper, 1983, Muenzer, 1986, Kurihara et al., 1992) Children are slow in reaching developmental milestones, and cognitive abilities in general. At some point, they reach a cognitive peak, and then begin to loose abilities. Young and Harper (1983) reported developmental regression beginning at age 6 1/2 on the average. The first sign of regression was apathy. They became progressively indifferent to their environment. They were less active, becoming nonambulatory at an average of 11.9 years of age. Speech was lost an average age of 10.9 years. At an average age of 12.4 patients " had eventually lost all skills and became totally bedridden and helpless". In cases of progressive terminal disease, teachers and collaborative teams can teach as many functional academics, daily living, and social skills as they can before children begin regressing.

Seizures are common in children with severe Hunter's syndrome (Young and Harper, 1983) and teacher need to recognize the various types of seizures. In their study, Young and Harper identified 17 out of 32 patients who had grand mal convulsions. The average age for seizures was 10 years. Medicine controlled the seizures. teachers can arrange the environment where students with a history of seizures won't hurt themselves (Lewis and Doorlag, 1995). They can educate classmates about seizures to reduce anxiety. Turning students on their sides during a seizure helps keep their airways clear. After seizures, teachers should check for breathing, record events, and follow school protocol.

Behavior

The literature describes children with severe Hunter's syndrome as having behavior problems (Young and Harper,1981, Muenzer, 1986). Young and Harper, 1981b) conducted a study of the psychosocial problems in Hunter's syndrome. Of 38 severely affected boys, 36 were described as having serious behavioral disturbances. The main problem parents reported was overactivity occurring from an average of 2 to 8 or 9 years of age. Four categories of behavior patterns were identified : overactivity, obstinacy, aggression, and exuberance. Overactivity was defined as " tireless, exhausting, indestructible and excitable"

Seventy six percent of the boys were described as overactive. Forty Seven percent were described as being obstinate. This was defined as " stubborn intractable behavior". Obstinate behavior included temper tantrums and throwing things. Forty two percent exhibited aggressive behavior. They were described as having anti-social behavior, and being violent, destructive or wild. Overenthusiasm or exuberance was reported in twenty-six percent of the boys. They were described as being helpful, playful, lovable and affectionate.

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Working with behavioral disordered children can be trying, even without physical disabilities.

Methods for working with students who have behavior disorders have been detailed in Kaplan and Carter's book suggesting a cognitive-behavioral approach to behavior management (1995). Some suggestions include conducting an ecological assessment to determine all the factors contributing to the undesirable behavior, making baseline assessments of behavior, giving parent behavior inventories, and carefully identifying the antecedents, operationally defined behavior(s) and the consequences of the behavior. This information would give teachers a place to begin modification of behaviors. Kaplan and Carter suggest identifying whether or not the behavior serves a function, like communication. The behavior then needs to be replaced by another appropriate behavior. Various types of reinforcement are recommended for good behaviors. Taking into consideration the poor expressive abilities of children with severe Hunter's syndrome (Young and Harper, 1983), communication seems like a reasonable cause for some of their inappropriate behaviors. A careful assessment of behavior would seem appropriate. Anti-social and aggressive behavior needs to be eliminated to increase social interaction. Studies show that positive social relations in young children influence other areas of learning including : intellectual, interpersonal, communicative and emotional development (Bates, 1975, Hartup, 1978, Parker & Asher, 1987, Rubin, 1980, cited in Salisbury, Gallucci, Palombaro, & Peck, 1985). Salisbury et al. (1995) describes five strategies that promote social relations among students who have disabilities and their typical peers. The five strategies are : active facilitation of social interaction, letting children solve problems alone or partially alone, exercises in empathy, modeling acceptance, and organizing the classroom structure to facilitate group activities. Strain and Kohler (1995) conducted a study of children with antisocial behaviors. Implementation of social skills training increased the target children's social interaction with typical peers significantly. Teachers of children with severe Hunter's can explore some of these strategies mentioned and incorporate them into their curriculum.

General Suggestions

Children with severe Hunter's syndrome present the educator and collaborative team with many physical challenges. Everyone who would be included in the children's education should be aware of the nature of the physical aspects of diseases like this and be prepared for any eventuality (Thomas, 1996). They must become familiar with daily medical procedures. Preventative care such as cutting up food into small bites, positioning children to facilitate breaking and prevent choking, encouraging range-of-motion exercises to slow muscle contractures and decrease muscle atrophy, are all aspects of education which should be considered when dealing with students like these. Teachers need to have good collaborative skills with other professionals like occupational therapists, speech and language pathologists, physical therapists, the school nurse, respiratory therapists, and others to provide an appropriate education and interaction with typical peers in the least restrictive environment (Lewis and Doorlag, 1995). Some of the most important members of the collaborative team for students with severe Hunter's are the parents.

Parent Collaboration

Parents are an integral part of the collaborating team for students with severe Hunter's syndrome (Simpson,1996). They are especially important in constructing the individualized education plan. They are valuable in the assessment of all areas of the child's education. They can work with the team in helping their children meet educational goals. Many times they must advocate for the needs of their children. They know their child better than anyone else and can provide helpful insight on what works best of their child. Simpson suggests that in order for parents to function to the best of their abilities, teachers may need to educate them on advocacy, teach them strategies and techniques for educating their child at home, provide resources that they may not be aware of, and provide understanding and compassion in times of transition.

Good parent-teacher collaboration is essential to a successful educational experience for children with severe Hunter's syndrome.

Providing Support for Parents

A disease like severe Hunter's syndrome can have devastating effects on the dynamics of a family. Young and Harper (1981) described some of the family problems that arose as a result of their children's disease. Almost all mothers of the 21 families felt a great deal of guilt. Many of them were not aware of the hereditary factor of the disease. Many could not accept the extent of the disability and held unrealistic views of their son's future achievements. Most mothers had reached a level of acceptance. The process suggested by Young and Harper was considered by Healy (1996). Healy points out stages of adjustment that parents go through when they learn that their child has a disability. He compares it to learning about the death of a loved one. Six stages of adjustment were described: initial shock and dejection; denial and avoidance; inward or outward feelings of anger; resignation, depression, guilt, shame, hopelessness, anxiety, and isolation; acceptance; and objective discussion of the child and advocacy issues. Teachers need to be aware of these stages and assist parents in the transitions. Healy suggests that teachers refer parents to support groups and counseling. He writes that sometimes teachers are the only people who can validate the parent's feelings. Parents can participate fully in their child's education when they have reached a level of acceptance. Until then, teachers can be patient and lend a sympathetic ear. In addition to empathy, teachers can provide parents with additional resources for support.

Teachers should be aware of the types of programs available as resources for families with disabled children
(Simpson, 1996). Becoming aware of a family's need for support is a first step to meeting the needs of the family. Agosta and Melda (1996) suggest the following objectives for supports and services needed by families; support families in their efforts to raise children at home; strengthen the role of the family in providing primary care, prevention of out-of-home placement and maintain family unity. These supports and services might include goods, services, and financial support. There are many available services that have been made available in recent years as a result of the legislation for individuals with disabilities according to Agosta and Melda. Some states have begun programs to help families with various needs resulting from the identification of a disabled child.

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The accessibility of these services by the family is different for different programs in different states. Simpson (1986) suggests that teachers make lists of state and local resources that are available for parents.

Teachers can help families identify their needs and direct them to appropriate services. Another consideration teachers can be aware of is the way these programs are arranged.

The way various family-oriented human service programs can be arranged was the focus of a study by Trivette, Dunst, Boyd and Hamby (1995). Trivette et al. identified four paradigms for differentiating between these programs; in professionally-centered models, professionals are considered to be the experts who determine the needs of the child and the family' family-allied models utilize the family in the interventions that the professionals deem appropriate; family-focused models consider families to be consumers of services, and thus assist families in making choices among options deemed appropriate by professionals; family centered models view professionals as instruments for families. Family-centered models with frequent contact from help givers who used empowering help-giving practice resulted in greater indication of personal control by family members. Agosta and Melda (1995) also recommended family-centered models for services. They suggest that parents need to feel empowered and in control of decisions that affect them. Family centered human service programs appear to empower families. Parents can learn to advocate for family needs and teachers are mandated to inform them of their legal rights in the education of their children
(Simpson, 1996).

Teachers of children with disabilities like severe Hunter's syndrome need to be aware of legislation that pertains to the rights of families regarding the education of their disabled child (Simpson, 1996). The Individual with Disabilities Education Act (PL 101-476) provides families with a number of rights. This document needs to be available for reference by teachers. It includes the following provisions for families :
(a) identification and provision of a free and appropriate education for all children,
(b) assessment that is nondiscriminatory and individualized,
(c) protection from inaccurate placement or denial or equal education and protection through due process,
(d) placement of students in the least restrictive environment
(e) Individualized Education Plans (IEPs) which ensure individualized appropriate education,and
(f) opportunity for parents to participate in their child's education

Teachers are required under the IDEA to inform parents in their native language or a language they understand of their rights of due process. Simpson suggests that teachers use understandable language when informing parents of their rights of due process.

Due process under the IDEA includes the right of parents to have access to their children's records (Simpson, 1996). Parents have the right to have their children evaluated by an independent person if they feel their child was not given an appropriate evaluation. They must be given written notice of any change in the placement or services for their child. This notice must be in a language that the parents can understand. Parents have the right to a surrogate parents If there is a conflict between the school and the parent that cannot be resolved, parents have the right to a fair and impartial hearing that is conducted by a neutral party. Teachers need to have access to the rights of due process so they can explain them to the parents.

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In addition to becoming sensitive to the needs of families who have children with disabilities, making resource information available, teaching advocacy and informing parents of their right of due process, teachers need to encourage parents to become involved in the educational plan of their children.

Simpson (1996) states that teachers of students with disabilities need to utilize parents when writing IEP's. In addition to the IDEA requirement of the rights of parents to participate in their child's education, Simpson suggests incorporating parents in all aspects of the educational process. This begins with parental input about the strengths and weaknesses of their children. Educational goals should be identified based on assessments including parent inventories and ecological assessment (Shea and Bauer, 1994) . Parents should inform teachers and other team members of their children's strengths and areas of weakness. Parents can work with team members to incorporate educational strategies at home. This will reinforce learning, and students will be more apt to generalize and maintain educational gains. When parents offer suggestions for the education of their children, and incorporate teaching strategies in their home, learning becomes more appropriate and functional.

Successful collaboration with parents is a vital part of the education of students with disabilities like severe Hunter's syndrome. Good teaching practice should not overlook the importance of ongoing communication and collaboration with parents.

Conclusion

Children with severe Hunter's syndrome present educators and collaborating team members with a number of considerations. The primary goal of educating these children is to provide them with as much independence as possible for the longest amount of time. Physical disabilities in affected children can be considerable and teachers need to be aware of possible problems. Learning about the nature of this disease and how it affects children will help educators to create an environment that is safe and strategies that will prevent further complications. Becoming aware of the behavior problems that sometimes occur in children with severe Hunter's syndrome can help teachers assess and prepare appropriate behavior interventions.

Sadly, the literature does not include many of the strengths that these children have. Parents of children with Hunter's syndrome often write to a publication about various topics of concern. Courage Magazine is a publication about various MPS diseases. Parents frequently refer to their affected children as being lovable, popular, coordinated in some sports, and good natured (1996,1997). Despite the failure of authors and researchers to address the strengths of these children, teachers and collaborating team members can, and should plan for their education on an individual basis. The IDEA provides a structure for the development of an educational plan that is based not only on the student's weaknesses but also on their strengths. Parents become especially vital in the development of the educational plan and can provide this type of information on an individual basis.

Parents can become more involved in their children's education when teachers assist them invalidating family concerns, offering resources for the family , informing them of their legal rights, and suggesting ways to become advocates for their children. Teachers need to be trained in collaboration techniques, become familiar with local resources for families, have access to and become familiar with the rights of due process as outlined in the IDEA, and facilitate parents in becoming advocates for themselves and their children.

References

Agosta, J. & Melda, K. (1995). Supporting families who provide care at home for children with disabilities. Exceptional Children, 62, 271-282.

Boice, C. & Boice, E. (1996) Courage. 32-33.

Healy, B. (1996). Helping parents deal with the fact that their child has a disability. CEC Today. 12-13.

Kaplan, J.S. & Carter, J. (1995). Beyond behavior modification : A cognitivebehavioral approach to behavior management in the schools. Austin, TX : PRO-ED Inc.

Kurihara, M.,& Kumagai, K. Goto, K. Imai, M. & Uagishita, S. (1992). Severe type Hunter's syndrome. Polysomnographic and neuropathological study. Neuropediatrics, 23, 269-256.

Lewis, R.B. & Doorlag, D.H. (1996). Teaching special students in the mainstream. (4 th ed.) Englewood Cliffs, New Jersey : Prentice-Hall.

Muenzer, J. (1986). Mucopolysaccharidosis. Advances in Pediatrics, 33, 269-302.

Mucopolysaccharide Disorders : Hunter Syndrome. Hicksville, New York : National MPS Society.

Polloway, E.A. & Patton, J.R. (1997). Strategies for teaching learners with special needs (6 th ed.). Upper Saddle River, New Jersey : Prentice Hall

Salisbury, C.L., Galluchi, C., Palombard, M.M., & Peck, C.A. (1995). Strategies that promote social relations among elementary students with and without disabilities in inclusive schools. Exceptional Children. 62, 125-137.

Shapirro, J., Strome, M. & Crocker, A.C. (1985) . Airway obstruction and sleep apnea in Hurler and Hunter's syndrome. Annual of OtolRhinolLarynsol 94, 458-461

Shaughnessy, M.F. (1996) . Working with multiply handicapped children. The Educational Forum, 61, 63-66.

Shea, T.M. & Bauer, A.M. (1994). Learners with disabilities : A social systems perspective of special education. Dubuque, IA : Wm C. Brown Communications

Simpson, R.L. (1996). Working with parents and families of exceptional children and youth. (3 rd Ed.) Austin, TX : Pro-Ed Inc.

Strain, P.S. & Kohler, F.W. (1995). Analyzing predictors of daily social skill performance. Behavioral Disorders, 21(1), 79-88.

Thomas, G.E. (1996). Teaching students with mental retardation : A life goal curriculum planning approach. Englewood Cliffs, New Jersey : Prentice Hall.

Trivette, C.M., Dunst, C.J., Boyd, K., & Hamby, D.W. (1995). Family oriented program models, help giving practices, and parental control appraisals. Exceptional Children, 62, (3), 271-282.

BEST COPY AVAILABLE

Wehneert, M., Hopwood, J.J., Schroder, W., & Hermann, F.H. (1992) . Structural gene aberations in mucopolysaccharidosis II (Hunter). Human Genetics, 89, 430-432.

Wraith, J.E. , Cooper, A., Thornley, M. Wilson, P.J.Nelson, P.V., Morris, C.P. & Hopwood, J.J. (1990). The clinical phenotype of two patients with a complete deletion of the iduronate-2-sulphatase gene (mucopolysaccharidosis II-Hunter syndrome). Human Genetics 87, 205-206.

Young, I.D. & Harper, P.S. (1981). Psychosocial problems in Hunter's syndrome. Child Care, Health, and Development, 7, 201-209.

Young, I.O., & Harper, P.S. (1983). The natural history of the severe form of Hunter's syndrome : A study based on 52 cases. Developmental Medicine and Child Neurology, 25, 481-489.

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